In the claims:

- 1-3. (Canceled)
- 4. (Withdrawn) An isolated protein encoded for by the nucleic acid sequence of SEQ ID NO:1.
- 5. (Withdrawn) An isolated protein of claim 4 having the amino acid sequence of SEQ ID NO:2.
- 6-10. (Canceled)
- 11. (Withdrawn) A method for treating an individual with basement membrane disease comprising administering an effective therapeutic amount of a protein of claim 4.
- 12. (Withdrawn) A method for treating an individual with basement membrane disease comprising administering an effective therapeutic amount of nucleic acid constructs containing an expressible nucleic acid sequence of SEQ ID NO:1.
- 13. (Withdrawn) A polyclonal antiserum containing antibodies specific for nephrin protein produced by immunizing an animal with a sufficient amount of the protein of claim 5 to stimulate an immune response.
- 14. (Withdrawn) A monoclonal antibody specific for nephrin produced by immunizing a rodent with a sufficient amount of the protein of claim 5 to stimulate an immune response.
- 15. (Withdrawn) A chimeric antibody comprising the variable domains of the antibody of claim 14 functionally attached to human antibody constant domains.
- 16. (Withdrawn) A kit for screening individuals for susceptibility to basement membrane disease, or the present of basement membrane disease, containing at least one antibody specific for nephrin.
- 17. (Withdrawn) A method for identifying a small molecule therapeutic for the treatment of proteinuria associated with kidney disease comprising screening candidate molecules for specific binding to the nephrin protein.
- 18. (Withdrawn) A method as in claim 17 wherein said specific binding effects a change in nephrin protein bioactivity.
- 19. (Previously presented) A method for diagnosing the presence of a basement membrane disease in an individual, comprising detecting the presence of a mutation in exon 2 or exon 26 of the NPHS1 gene encoded for by the nucleic acid sequence of SEQ

- ID NO:1, wherein the mutation in at least one of the exons results in a premature stop codon in the exon.
- 20. (Canceled)
- 21. (Previously presented) The method of claim 19, wherein the mutation in exon 2 comprises a two base pair deletion.
- 22. (Previously presented) The method of claim 21, wherein the NPHS1 gene is amplified prior to detecting the presence of the mutation in exon 2.
- 23. (Previously presented) The method of claim 22, wherein the amplification is by PCR and the primers used for amplification specifically amplify the exon 2 region of the NPHS1 gene.
- 24. (Previously presented) The method of claim 23, wherein the primers used for amplification comprise DNA sequences comprising SEQ ID NO:3 or SEQ ID NO:4.
- 25. (Previously presented) The method of claim 19, wherein the mutation in exon 26 comprises a single base change.
- 26. (Previously presented) The mutation of claim 25, wherein the single base pair change results in the nonsense mutation CGA->TGA.
- 27. (Previously presented) The method of claim 25, wherein the NPHS1 gene is amplified prior to detecting the presence of the mutation in exon 26.
- 28. (Previously presented) The method of claim 27, wherein the amplification is by PCR and the primers used for amplification specifically amplify the exon 26 region of the NPHS1 gene.
- 29. (Previously presented) The method of claim 28, wherein the primers used for amplification comprise DNA sequences comprising SEQ ID NO:5 or SEQ ID NO:6.
- 30. (Previously presented) The method of claim 29, wherein a novel restriction site is detected in the amplified product.
- 31. (Previously presented) The method of claim 30, wherein the novel restriction site is susceptible to digestion with DdeI.
- 32. (Previously presented) A method of determining whether an individual is at risk for developing a congenital nephrotic syndrome of the Finnish type, comprising analyzing a nucleic acid sample containing the NPHS1 gene encoded for by the nucleic acid sequence of SEQ ID NO:1, wherein the method comprises analyzing the exon 2 or

- exon 26 region of the NPHS1 gene, wherein an individual at risk for developing a congenital nephrotic syndrome has at least one mutation in either or both of the exons.
- 33. (Canceled)
- 34. (Previously presented) The method of claim 32, wherein the mutation in exon 2 comprises a two base pair detection deletion.
- 35. (Previously presented) The method of claim 34 wherein the NPHS1 gene is amplified prior to detecting the presence of the mutation in exon 2.
- 36. (Previously presented) The method of claim 35, wherein the amplification is by PCR and the primers used for amplification specifically amplify the exon 2 region of the NPHS1 gene.
- 37. (Previously presented) The method of claim 36, wherein the primers used for amplification comprise DNA sequences selected from the group consisting of SEQ ID NO:3 or SEQ ID NO:4.
- 38. (Previously presented) The method of claim 32, wherein the mutation in exon 26 comprises a single base pair change.
- 39. (Previously presented) The mutation of claim 38, wherein the single base pair change results in the nonsense mutation CGA->TGA.
- 40. (Previously presented) The method of claim 39, wherein the NPHS1 gene (SEQ ID NO:1) is amplified prior to detecting the presence of the mutation in exon 26.
- 41. (Previously presented) The method of claim 40, wherein the amplification is by PCR and the primers used for amplification specifically amplify the exon 26 region of the NPHS1 gene.
- 42. (Previously presented) The method of claim 41, wherein the primers used for amplification comprise DNA sequences selected from the group consisting of SEQ ID NO:5 and SEQ ID NO:6.
- 43. (Previously presented) The method of claim 42, wherein a novel restriction site is detected in the amplified product.
- 44. (Previously presented) The method of claim 43, wherein the novel restriction site is susceptible to digestion with DdeI.
- 45. (Currently amended) A method for determining that an individual is not at risk for developing congenital nephritic syndrome of the Finnish Type, wherein the

syndrome is associated with a mutation in exon 2 or exon 26 of the syndrome NPHS1 gene, comprising analyzing a nucleic acid sample containing the syndrome gene, wherein the method comprises analyzing the exon 2 or exon 26 region of the NPHS1 gene encoded for by the nucleic acid sequence of SEQ ID NO:1, wherein the individual not at risk for developing the syndrome does not have a mutation in exon 2 or exon 26.

46. (Canceled)

- 47. (Previously presented) The method of claim 45, wherein the NPHS1 gene is amplified prior to analysis.
- 48. (Previously presented) The method of claim 47, wherein the amplification is PCR amplification using primers comprising a DNA sequence selected from the group consisting of: SEQ ID NO:3, SEQ ID NO:4, SEQ ID NO:5, and SEQ ID NO:6.
- 49. (Previously presented) A method for detecting the presence or absence of a mutation in the NPHS1 gene, comprising the steps of:

analyzing a nucleic acid test sample containing the NPHS1 gene encoded for by the nucleic acid sequence of SEQ ID NO:1 for at least one mutation in exon 2 or exon 26 of the gene;

comparing the results of the analysis of the test sample of step a) with the results of the analysis of a control sample, wherein the control sample comprises a NPHS1 gene encoded for by the nucleic acid sequence of SEQ ID NO:1 without a mutation in exon 2 or exon 26; and

determining the presence or absence of at least one mutation in exon 2 or exon 26 in the test sample.

- 50. (Canceled)
- 51. (Previously presented) The method of claim 49, wherein the mutation in exon 2 is a two base pair deletion and the mutation in exon 26 is a single base pair change, wherein either mutation results in a premature stop codon in the exon.
- 52. (Previously presented) The method of claim 49, wherein the NPHS1 gene is amplified prior to analysis.
- 53. (Previously presented) The method of claim 52, wherein the amplification is PCR amplification using primers comprising a DNA sequence selected from the group consisting of: SEQ ID NO:3, SEQ ID NO:4, SEQ ID NO:5, and SEQ ID NO:6.

54. (Withdrawn) A primer comprising a nucleic acid sequence comprising SEQ ID NO:3, SEQ ID NO:4, SEQ ID NO:5, or SEQ ID NO:6.